

PICK'S DISEASE

(MEDIASTINO-PERICARDITIC PSEUDO-CIRRHOSIS OF THE LIVER)

A CASE, WITH PERICARDIAL RESECTION AND RECOVERY

BY G. F. STRONG, M.D.

Vancouver

WHILE adhesive pericarditis is not an uncommon finding at autopsy, true Pick's disease is comparatively rare. A satisfactory outcome after a total period of three years' observation, including surgical treatment, would seem to justify the following report.

E.B., male, aged 18, was first seen in March, 1935, suffering from acute pericarditis of two months' duration. The present illness had developed in January, 1935, as an acute febrile illness associated with sore throat, malaise, night sweats, loss of strength; then epigastric and pericardial pain or soreness appeared. This pain was not related to food, but was made worse by deep respiration or cough. He was admitted to the Vancouver General Hospital on February 7, 1935. During the month in hospital he was acutely ill, with a daily temperature of 99 to 102°.

Past history.—He was born in England; was well as a child, with only the usual diseases, measles, mumps, whooping-cough; no scarlet fever or diphtheria. He came to Canada in 1919. There was no history of previous rheumatic infection, except that in 1934, following a camping trip, he had had some rheumatic pains and was kept in bed for two weeks. He did not see a doctor, and recovered and returned to his work in a door factory.

Family history.—Essentially negative.

Physical examination.—The patient was a well developed rather poorly nourished boy of the stated age. He appeared acutely ill and was slightly orthopneic. Pulse 110, regular; temperature 100°; blood pressure 110/70. The heart was enlarged to percussion; the sounds were weak and distant; no murmurs; no rub. The lungs were clear except for dullness at the left base, with diminished tactile and vocal fremitus and breath sounds. The abdomen was flat; no ascites. The liver and spleen were not palpable. There was no peripheral oedema; no distension of cervical veins. The urine was normal. Blood count: red blood cells 3,800,000; hgb. 78 per cent; white blood cells 6,675; polymorphonuclears 70 per cent; lymphocytes 23 per cent; mononuclears 4 per cent; eosinophiles 2 per cent. X-ray showed pericardial effusion. Electrocardiogram: T 1 and 2 inverted, T 3 flat (pericardial tamponade).

Diagnosis.—Acute pericarditis and left-sided pleurisy with effusion.

Progress notes.—March 20th, aspiration of pericardium attempted but not successful. April; x-ray, left pleural effusion. Pleural paracentesis on two occasions (fluid sterile). Inoculation of a guinea pig was negative for tuberculosis. May; continuous temperature of 99 to 102°. First appearance of ascites and some slight oedema of ankles. The urine output diminished, fifteen to twenty ounces per day. Urinalysis was normal. June; relapse. Hæmoptysis followed aspiration of the chest. July; he was given ammonium chloride and salyrgan for the ascites with good effect. August to December; gradually up and about with an occasional mild febrile relapse. Repeated mercurial diuretics and occasional aspiration, at first of the chest and abdomen, then only of the abdomen. He was discharged from hospital on December 23, 1935.

During 1936 this patient was followed in the outpatients' department, where he was treated with tonic doses of digitalis (grs. 1½ per day), with iron for a recurring secondary anaemia, and diuretics. Ammonium chloride and salyrgan, or salyrgan alone, were given in repeated attempts to control the ascites. In spite of this treatment and rigid limitation of the fluid intake to thirty ounces in the twenty-four hours it was necessary at irregular but frequent intervals to resort to abdominal paracentesis: 130 to 326 ounces of slightly turbid, straw-coloured fluid were removed each time. The patient's weight varied from 130 to 158 lbs., depending on the amount of dropsy present. While some oedema of the ankles did occur from time to time it was very slight in proportion to the ascites. A scoliosis of the dorsal spine developed during the latter part of 1936. The venous pressure was persistently high, varying from 26 to 29 cm. of water. The urine output continued at about 15 ounces in twenty-four hours, except after salyrgan, when a considerable diuresis up to 100 to 120 ounces would occur.

In the spring of 1937 it was decided to resort to surgery as his condition seemed to have become stationary and the constantly recurring ascites was a great handicap. In February, 1937, x-ray studies were undertaken to confirm the diagnosis of adhesive mediastino-pericarditis. Films taken in various positions failed to reveal the normal shift in the position of the heart or the mediastinum. Kymographic studies were attempted, but were unsatisfactory because the borders of the heart were obscured by bilateral pleural thickening.

In June the patient was readmitted to hospital for further study as a preliminary to operation. Physical examination revealed a pulse of 80, blood pressure 110/80, and a weight of 156 lbs. There was evidence of mediastinal block, with persistently high venous pressure, engorgement of the neck veins, and recurring ascites, slight enlargement of the liver, an apparently small (palpation) fixed heart, dullness at the bases of both lungs especially the left, and some slight oedema of the shins. The urine was normal and renal function tests were satisfactory. Blood count: red blood cells 4,300,000; hgb. 68 per cent; white blood cells 4,700; differential count was normal. Total serum protein was 7; serum albumin 2.8; serum globulin 4.2; blood calcium 10.5. The blood sedimentation rate was 22 mm. in 45 minutes. The venous pressure on June 6th was 29 cm. of water. Electrocardiogram showed slight change since that taken in 1935—T 1 up, T 2 flat, T 3 inverted; low potential. X-ray: small fixed heart, immobile mediastinum, increased density in both bases. Abdominal paracentesis was done on June 3rd.

June 8th.—A pericardiotomy was performed by Dr. A. B. Schinbein, who made the following notes regarding the operation.

"The approach to the pericardium was made by a curved incision, with the convexity to the right, starting in the mid-sternal line opposite the second costal cartilage and ending at the mid-sternum opposite the sixth costal cartilage. This flap was raised; the costal cartilages of the third, fourth and fifth ribs were removed. The internal mammary artery was tied. One inch of the third, fourth and fifth ribs was removed. The pleura was separated by blunt dissection and retracted. One-half of the sternum from the

third to the fifth ribs was removed. The parietal pericardium was thus freely exposed. The pericardium was thickened, but there were no evidences of calcification. An opening was made into the pericardium and adhesions between the heart and pericardium were found. The heart appeared to be normal in size; it was certainly not enlarged. The adhesions over the apex and the right ventricle were not dense and were easily separated. As one approached the auricle, however, the adhesions became more dense and the wall of the auricle was quite thin. Separation here was done by blunt and sharp dissection, as far as possible without exposing the opposite side. The parietal pericardium was removed over this whole area. The edge of the pericardium was sutured to the pectoral muscle over the ends of the ribs by interrupted sutures. The skin incision was closed without drainage."

The post-operative course was surprisingly easy. The patient had been kept in a semi-reclining position during the operation and that position was maintained for several days. He was kept in an oxygen tent for twenty-four hours after the operation.

After three weeks he was allowed up. The venous pressure did not change, and readings after the operation were as follows: June 9th, 25 cm.; June 11th, 21 cm.; June 28th, 26 cm.; July 21st, 23 cm. Ascites recurred after the operation and abdominal paracentesis was done three times on July 2nd (153 ozs.), July 17th (178 ozs.), August 26th (165 ozs.). July 19th, complete blood count showed: red blood cells 3,600,000; hgb. 75 per cent; white blood cells, 6,750; with normal differential. The electrocardiogram showed some improvement in amplitude of Q.R.S. complex.

The patient was discharged from hospital on August 28, 1937, and treatment was continued through the out-patients' department. During September the average daily urine output was 15 to 20 ounces, except in response to the intravenous injection of 2 c.c. of salyrgan. No abdominal paracentesis was done after patient left hospital. Other treatment at this time consisted of digitalis, grs. $1\frac{1}{2}$ per day, for relief of tachycardia and slight dyspnoea, and iron for the anaemia.

Quite suddenly, in October, four months after operation, the patient noted an increase in urine output to 50 ounces in twenty-four hours. The use of salyrgan was thereupon discontinued and no further diuretics have been necessary. There has been no return of the ascites to the present time (March, 1938) and there is only very slight pitting of the shins at the end of the day.

The only discomfort the patient suffers at present is dyspnoea. Recent examination revealed the fact that he is definitely improved. He now weighs 157 lbs., and since there is practically no dropsy this represents an actual gain in weight. Pulse continues slightly fast, 100 and regular; blood pressure 120/80. The heart sounds are of fair quality; no murmurs. There are dullness and suppression of breath sounds at the bases of both lungs, more especially on the left. The abdomen shows no free fluid, and the liver is not now palpable. There is slight pitting of the shins. There is a scoliosis of the dorsal spine with a concavity to the left. There is still engorgement of the neck veins, accentuated when the patient lies down, at which time his face is suffused. The venous pressure remains high—26 cm. of water. Blood non-protein nitrogen is 34 mg. per 100 c.c. Total proteins 6.2, albumin 3.4, globulin 2.8. Urinalysis is normal and the total twenty-four hours' output is about 50 ounces.

DISCUSSION

The cause of this condition is not entirely clear. White¹ in 1931 mentioned rheumatic infection as the commonest cause, with tuberculosis

next. In the St. Cyres Lectures at the National Heart Hospital in London in 1935,² however, he reported 15 cases, 2 due to tuberculosis, 3 due to other infections, none due to rheumatic infection, and 10 due to unknown causes. The present case would appear to be due to a rheumatic type of infection. Tuberculosis seems to have been definitely excluded.

The reversal of the normal serum-albumin-globulin ratio has been reported in other cases and is probably only an evidence of the albumin loss due to the repeated aspirations. The fact that the ratio returned to a nearly normal reading five months after aspirations were discontinued would confirm this view. It is possible that this reversal of albumin-globulin ratio indicates some damage to the liver cells. No other tests of hepatic function were attempted in this case.

The observation that the urine output suddenly and apparently spontaneously increased four months after the operation, with the concomitant disappearance of the ascites, is also not new. Apparently at this time the back pressure in the hepatic veins had subsided sufficiently to relieve the hepatic congestion causing the portal obstruction. There is still in this patient some evidence of interference with the emptying of the superior vena cava. We feel that this is due to the fact that dense adhesions around the auricles were not entirely separated.

There must be differentiation between polyserositis (Concato's disease), and mediastinopericarditic pseudo-cirrhosis of the liver (Pick's disease). The first may lead to the second. On the other hand, the second may occur without polyserositis, and *vice versa*. The outstanding feature of Pick's disease is the evidence of constriction of the heart and of the great veins near the heart. Diastolic filling is impaired and over-filling of the great venous trunks is produced. The hepatic engorgement produces a pseudo-cirrhosis of which ascites is the prominent feature. Signs of partial obstruction of the superior vena cava are increased engorgement of the neck veins, and suffusion and swelling of the face, the so-called upper mediastinal syndrome. In this case we feel that greater success might have been achieved had we been able to free more of the adhesions over the right auricle. The density of the adhesions encountered over the very thin walled auricles made us unwilling to risk further surgical interference.

It is always difficult to decide when to operate. Obviously it is dangerous to operate too soon after the initial infection. On the other hand to wait too long is to invite permanent changes in the liver which might persist after the operation. While this young man is not

entirely well, he is immeasurably better than before his operation, and now presents no evidences of portal obstruction.

REFERENCES

1. WHITE, P. D.: *Heart Disease*, Macmillan, Toronto, 1931, p. 517.
2. *Idem*: Chronic constrictive pericarditis, *The Lancet*, 1935, 2: 539 and 597.

THE PERFECTING OF ANÆSTHESIA*

BY WESLEY BOURNE

Montreal

THE perfect anæsthetic is far to seek, and, vague-looming, may remain in the realm of the ideal. It is difficult to conceive of a chemical entity capable of producing narcosis without untoward action and at the same time suitable for all manner and conditions of men. Rather is it better to believe that our end and aim in ease will be attained the sooner by using such anæsthetics as are least harmful, by improving the methods of their administration, and by being prepared to offset or lessen deleterious effects.

Mythical studies and perusals of the stories of our earlier heroes alike reveal that the germ of the idea of surcease from suffering is coeval with the dawn of creative thought. Despite the stirring nature of the many incidents recounted of superstitions, rites and ceremonies in the one, and of mysticisms from the other; notwithstanding the pleasurable illusiveness to be found in both, they may not be iterated at this juncture. Following a series of brilliant discoveries in chemistry during the latter part of the eighteenth century and the early half of the nineteenth, anæsthesia as we understand it today had its inception. The lives of Humphry Davy (1798), Michael Faraday (1818), Henry Hill Hickman (1828) — the most neglected — Crawford W. Long (1842), Horace Wells (1844), William T. G. Morton (1846), James Young Simpson (1847), John Snow (1848) — the most erudite — and some others; the lives of these men all remind us of the early achievements in modern anæsthesia, and are filled with such human and emotional appeal as to make the story one of the most interesting in the development of medical science. Thus, since the forties of the late century, nitrous oxide, ethyl ether and chloroform have been used, their beneficial and in-

jurious results have been studied, and means have been devised to counteract or alleviate the latter. Further, many other chemical substances have been investigated, several of which are useful as anæsthetics or aids in anæsthesia, so that there are a number of drugs from which to choose for administration alone, in combinations or sequences. Concomitantly, a variety of methods for their employment are being developed. Both methods and materials may be suited to the general condition as well as the special requirements of a given individual.

Just as pharmacology has been defined as the study of the action of chemical substances upon living things, so too, the ancillary topic, anæsthesia, has to do with the effects of anæsthetics on single cells as well as on highly complex organisms. But more than this is implied, for a good anæsthetist must be artistic, indeed, æsthetically so. Thoroughly he must understand the many physical phases of his work and be at the same time a metaphysician, quietly perceiving the psychisms of his immediate environment. This view might be pronounced an aphorism in the principles of anæsthesia.

As pharmacologists are men of discrimination and erudition, new drugs are not brought from the laboratory until very careful studies have been made of their effects on lower forms of life, and often on the investigator himself. Thus it is that an anæsthetic is not acceptable for general use on the human being before the completion of extremely thorough experiments.

Anæsthesia is divided into two main branches, namely, regional anæsthesia and general anæsthesia. Concerning the former, it is sufficient, at this time, to say that sensation may be suspended in any given region of the body by topical application or injection of such drugs as cocaine, procaine and percaine. It is in this system of anæsthesia that one encounters the

* From a lecture delivered at a meeting of the McGill Chemical Society, February 4, 1938.